A CASE OF MUSCULAR DYSTROPHY.

(Landouzy-Dejerine.)

By Charles Gilbert Chaddock, M.D.

PROFESSOR OF DISEASES OF THE NERVOUS SYSTEM, MARION-SIMS COLLEGE OF MEDICINE, ST. LOUIS, MO.

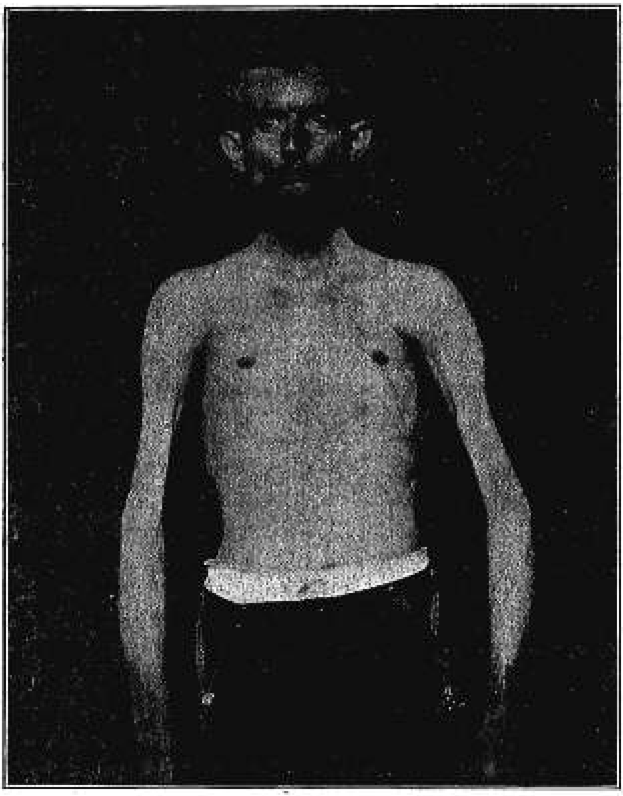
The following case is worthy of record owing to the comparative rarity of this type of myopathic muscular atrophy.

The patient is a man, single, 39 years old, a native of St. Louis, but of foreign-born parents; by occupation a machinist. He followed his trade during many years until increased feebleness rendered it impossible for him to continue work. No history of his father's family is obtainable, but his father died at the age of 61 of some unknown disease; he was a drinker, given to excess. The mother is living, and 65 years old; the history of her ancestry is negative. There is no history of nervous trouble either in the father or the mother. The patient had three brothers, two of whom died in early childhood, the third died at the age of 18 of some acute disease; a sister died in childhood. The history of the health of these members of the family is negative. No paralysis or atrophy was ever observed in any of them.

In early childhood, the patient had an acute inflammatory disease of the lungs, but from the date of this illness until manhood he had no severe sickness of any kind. During his childhood he was active and agile and able to engage in the ordinary sports of boyhood. However, he states that he early tried to whistle, but that he was never able to acquire the accomplishment. His lips refused to act. At the age of 15 a bit of emery got into his left eye, and the eye was neglected for some time owing to his unwillingness to have it treated. From this accident he dates his recognition of his inability to completely close his eyelids. It is probable that this was of much earlier origin, and the accident served to call his attention to it. His present peculiarities of countenance, he says, he has had since he can remember; the depressions in his cheeks have always been noticeable, though more marked of late years. At the age of 24 he remarked a growing weakness in his right arm, affecting especially the movements of extension and flexion of the forearm. Later he noticed a similar weakness of the left arm. At the age of 27 he noticed some peculiarity of his attitude in standing and of his movements in walking. He found that at his work he was resting his weight unconsciously on the balls of the feet, so that callosities were developed beneath the great toes, and became more or less painful and troublesome. He states that weakness has been always most apparent on the right side, a difference which he attributes to his greater use of that side. He gives no history of pain or of disturbances of sensation, except that in his right arm, above the elbow and throughout the muscles of that arm, he experienced aching and throbbing after having worked too long. This sometimes was troublesome and disturbed his rest. His greatest weight was at the age of 22—148 pounds.

The weakness noticed in early manhood gradually increased until he reached his present condition, which has kept him from active work for some years. He has attempted to perfect himself in writing, hoping to be able to employ himself in that way; but he met an obstacle in the cramping of his fingers when he continued the effort to write. He gives no history of ever having observed or felt fibrillary twitching in any of his muscles, and it may be added that repeated observation of him during several weeks has never revealed any such phenomenon. There has never been at any time vesical disturbance, and there has been no alteration of sexual power. The deep reflexes at the knees, the reflex of the tendo-Achillis and those in the arms, are present, but much diminished. The jaw-jerk is also present. The muscular power of the lower extremities is remarkably good for all movements, considering the evident amount of atrophy of the muscles. The grip is powerful, but stronger on the left than on the right side. Movements of the arm, especially those of extension and flexion of the forearm, and movements of the arm at the shoulder, are much weakened, though there is no actual loss of any single movement. There is neither objective nor subjective disturbance of sensibility.

An examination of the muscles of the body in detail reveals the following conditions, which are more or less clearly illustrated by the accompanying photograph: The skin of the brow is smooth, devoid of all the usual lines, and the patient cannot wrinkle the brow, though he can move the skin of the brow by contracting the occipito-frontalis and the orbicular muscles of the eyes alternately. He is unable to completely close the eyes, and his power to resist an effort to open them is much diminished. The natural form of the nares is altered so that the openings are mere slits, owing to the atrophy and weakness of the nasal muscles. The buccinator muscles are so reduced that the cheeks are depressed in deep indentations beneath the cheek bones. The corners of the mouth cannot be raised; the mouth is prominent and the lips turn outward in a characteristic fashion (*pot de chambre*). The smile is transverse. There is no weakness of the essential muscles of mastication, which causes a prominence at the angles of the jaw. The tongue shows no atrophy and is normally protruded, but it is in constant vermicular



movement. The pupils react normally to light and in accommodation, and there is no evidence of weakness or paralysis in the external muscles of the eye. The edge of each cornea presents a distinct cloudy circle, which is identical with the so-called arcus senilis. It may be added that the patient has worn glasses for presbyopia for six years. The muscles of the neck, while diminished in volume, have still much force, and are very firm to the touch. The muscles of the shoulder girdle are greatly atrophied. All these muscles are involved except the infraspinatus. The fibers of the pectoral muscles are scarcely perceptible, and at the seat of the supraspinatus there is a deep hollow on each side. The deltoids have almost disappeared over the points of the shoulder, but they present a characteristic ball-like prominence in the middle, which is evident in the photograph. The trapezius muscles are much reduced in volume, and also the rhomboids and the serrati, causing the characteristic wing-like projection of the scapulæ, with elevation of their superior angles. Other muscles of the back show no marked atrophy, and the patient complains of no weakness of the back. The abdominal muscles are in good condition. The biceps, coraco-brachialis, brachialis anticus, and triceps are reduced to thin bands on each side, but when the biceps contracts there is noticeable in its middle third a ball-like prominence, which shows that the active fibers in the muscles are those of its middle third. This phenomenon is observable on both arms. The contrast between the muscles of the arm and those of the forearm is very striking. The forearms are reduced in volume, but the muscles are abnormally hard, and their force, as shown by the grip and the power to resist extension of the hand on the wrist is, if not normal, at least up to what might be expected of a forearm of similar size. The intrinsic muscles of the hand are somewhat affected. This is true especially of the hypothenar and thenar muscles, and there is evidence of slight atrophy of the adductor pollicis and interrossei. The atrophy of the intrinsic muscles of the hand is more marked on the right side. The muscles about the pelvis are reduced in volume but have practically normal force. The atrophy of the muscles of the thighs is most marked in the lower third. The quadriceps extensor cruris being the muscle most markedly implicated. In the leg, the calf muscles are in fairly good condition, with some tendency to contracture, but the anterior tibial muscles are much reduced in volume. The intrinsic muscles of the feet have not suffered decidedly. There is toe-drop on the left side in walking. All the muscles react to faradism, even those most atrophied, though for the latter a more powerful current is necessary to induce contraction.

It is remarkable that the patient is still capable of almost all ordinary movements, notwithstanding the great wasting of so many muscles. He suffers more from weakness of movement than from actual loss of movement. The strength of his hands and forearms is such that he indignantly repels any suggestion that he is weakened in this respect.

The most remarkable condition observable, aside from the muscular atrophy, is the hard and wooden-like consistency of the muscles that have thus far escaped marked atrophy. They do not give the sensation of normal muscle, but rather that of a muscle in intense contraction, even when they are at rest.

Another point of interest is the entire absence of subcutaneous fat over the whole body, so that the patient has the appearance of being reduced to skin and bones.

This wasting of subcutaneous tissue, contemporaneous with muscular atrophy, and the evident abnormal condition of the muscles which are not yet advanced in atrophy, are worthy of special remark.

In the history of the development of this condition, it may be noted that undoubtedly the beginning of the trouble was in the face, that it subsequently attacked the arms, and later the lower extremities became involved, and finally the hands have been invaded. Even at the present time, with atrophy far advanced and quite generalized, the wasting at the roots of the extremities predominates decidedly over that at the distal extremities of the members. The characteristic prominence of the middle third of the wasted deltoids is also worthy of note. The absence of all fibrillary contractions, as well as the absence of all sensory and central disturbances are points which render a diagnosis of myopathic atrophy certain, and the order in which the atrophy has manifested itself places the case among those first described by Landouzy and Dejerine.

In this case, though there is no evidence of atheromatous changes in the vascular system, it is worth noting that there is some evidence of early senility in the condition of the cornea and the necessity for glasses to correct a presbyopia which began at the age of 33.